Case Report: Trabecular juvenile ossifying fibroma presenting as a sellar mass

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Abstract: A 15-year-old girl presented with left occulomotor nerve palsy and was found to have a space occupying lesion of the sellar region with invasion of the left cavernous sinus. A transsphenoidal approach lead to subtotal removal of a solid tumor with some remnants in the cavernous sinus and revealed the diagnosis of trabecular juvenile ossifying fibroma (JOF). A repeat magnetic resonance imaging was obtained within 1 month that showed intrasellar recurrence and growing tumor in the cavernous sinus. Therefore, a combined transsphenoidal and transcranial approach was performed to more aggressively remove the tumor. Subsequently, adjuvant proton radiotherapy was performed. JOF of the trabecular type is a rare fibro-osseous lesion of the craniofacial skeleton almost exclusively occurring in the maxilla or the mandible. To our knowledge, this is the first case of this tumor entity presenting as a sellar mass.

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Case report:

Trabecular juvenile ossifying fibroma presenting as a sellar mass

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Introduction

Juvenile ossifying fibroma (JOF) is a rare fibro-osseous lesion of the craniofacial skeleton occurring predominantly in children [1]. Two separate entities, psammomatoid juvenile ossifying fibroma (PsJOF) and trabecular juvenile ossifying fibroma (TrJOF) are described in the literature [2]. Microscopically, the tumor consists of a cellular fibrous stroma and of a mineralized osseous component in either a psammomatoid or a trabecular pattern [3]. PsJOF is the more frequently reported subtype and involves primarily the bones of the paranasal sinuses and of the orbit [4-5]. In contrast, TrJOF almost exclusively affects the mandible or the maxilla, tends to occur in younger patients and is characterized by a less favorable clinical behavior with a locally aggressive growth pattern and a high rate of recurrence [2]. Therefore, it is also termed juvenile aggressive fibroma. Usually, TrJOF presents as a painless swelling of the jaw bones and leads to facial asymmetry [6]. Surgery is the mainstay of therapy, but due to the infiltration of the surrounding tissue, the tumor often cannot be shelled out in its entirety and has a high rate of recurrence. To our knowledge, we describe the first case of TrJOF of the sella turcica.

Case Report

A 15 year old female presented with a 4 week history of diplopia, anisocoria (mild mydriasis on the left), mild ptosis on the left eye, and frontal pressure-type headaches. She was initially seen by an ophthalmologist who referred her with the diagnosis of left occulomotor nerve palsy to the Children's Hospital for further examination. Preoperative imaging showed a contrast-enhancing space occupying lesion (4x3x2cm) of the sellar region with extension into the cavernous sinus bilaterally and compression of the optic chiasm (figure 1). The sphenoid sinus was filled with tumor and the bony structures of the dorsum sellae and the superior aspect of the clivus were infiltrated by the lesion. Endocrinologic work-up revealed partial hypogonadotropic hypogonadism with primary amenorrhea and pubertas tarda, but no inadequately elevated hormone levels. Initially, a non-secretory macroadenoma of the pituitary gland was suspected and a transsphenoidal resection with intra-OP MR imaging (ioMRI) (PoleStar N20, 0.15 T, Medtronic Navigation) was performed. Intraoperatively, this solid lesion appeared white-grayish and had a rough cartilaginous consistency. Tissue samples from this procedure, as decribed below in more detail, were classified as trabecular juvenile ossifying fibroma. Definitive diagnosis was delayed due to the request for second and third opinions from external pathologists. Four weeks after the first resection follow-up MRI showed residual tumor (figure 3) and a second procedure using a combined transsphenoidal and pterional approach with ioMRI was performed (figure 2). This time, a nearly gross total resection has been accomplished as shown by the follow-up MRI scans (figure 3). The postoperative course was uneventful and the patient's symptoms (especially anisocoria, ptosis and headaches) started to improve during the first post-operative weeks. In accordance with the management recommendations by the hospital's interdisciplinary neurooncology tumor board, the patient underwent proton radiotherapy at the Swiss Paul Scherrer Institute. A total dose of 59.4 Gy(RBE) was delivered postoperatively in 33 daily treatment fractions. Double vision improved significantly after

proton radiotherapy. Clinical follow-up and re-imaging (figure 3) 12 months after her initial presentation revealed largely resolved ocular symptoms, unimpaired olfaction, and no headaches. Her endocrinologic follow-up showed the hormone levels seen in table 1. She reports no new symptoms and is participating in school with no problems.

Histology

Histological examination revealed a cell-rich neoplasm consisting predominantly of fibroblastic cells. The cells were located in a partly fibrous, partly compact-appearing, eosinophilic matrix (figures 4 and 5). Elastica van Gieson staining detected a net-like structure (figure 6), which became compact in some areas forming immature osteoid. The neoplastic cells were spindle-shaped to oval and some giant cells could be found. Immunohistochemical staining of the cells and the matrix surrounding the trabecular structures was positive for Procollagen 1. Additionally, the cells stained positive for Osterix, a transcription factor known to be expressed in osteoblasts.

The typical histological features of trabecular juvenile ossifying fibroma are aggregates of giant cells in a cell-rich fibrous stroma containing bundles of cellular osteoid and bone trabeculae without osteoblastic rimming [7].

Discussion

Although TrJOF represents a rare fibro-osseous lesion, early management decisions anticipating the clinical behavior of those lesions should be made, for they might be histologically benign, but locally aggressive as in this case. For that reason, most aggressive tumor removal as soon as possible is critical. In addition, this case also highlights the suitability of ioMRI since the degree of resection is of major importance, especially if the tumor infiltrates delicate structures like the cavernous sinus. The advantages of ioMRI were already addressed in other tumor entities [8-9] and also showed a benefit in the grade of resection in this patient.

Radiation therapy is generally not employed in the primary management of these essentially non-malignant lesions for justified concerns of late damaging effects to developing normal tissues and induction of malignancy later in life. However, precision radiotherapy has been used for a variety of benign, but locally aggressive mesenchymal tumors in anatomic locations precluding complete, microscopic surgical resection and following recurrent growth [10]. Proton Radiotherapy has demonstrated its advantage of precise tumor targeting combined with maximum normal tissue sparing and is an accepted and often preferred radiotherapy modality in the treatment of pediatric, solid tumors [11]. Proton Radiotherapy permits delivery of higher radiation doses compared to conventional photon radiotherapy [12] . This maximizes chances for permanent local control given the difficulties of salvage surgery in case of recurrence in the same location.

Conclusion

Trabecular juvenile ossifying fibroma is a locally-aggressive juvenile tumor mostly found extracranially. This is the first case of TrJOF mimicking a macroadenoma of the pituitary. Even though patients with such a diagnosis are rare, one should always consider a fibro-osseous lesion when dealing with a sellar mass. Treatment strategies should be interdisciplinary and should include an early, aggressive tumor removal. In this setting, ioMRI is a helpful tool to achieve a high degree of resection. Due to its precise tumor targeting, proton radiotherapy is an excellent modality in the treatment of these locally-challenging pediatric tumors.

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FIGURES and TABLES

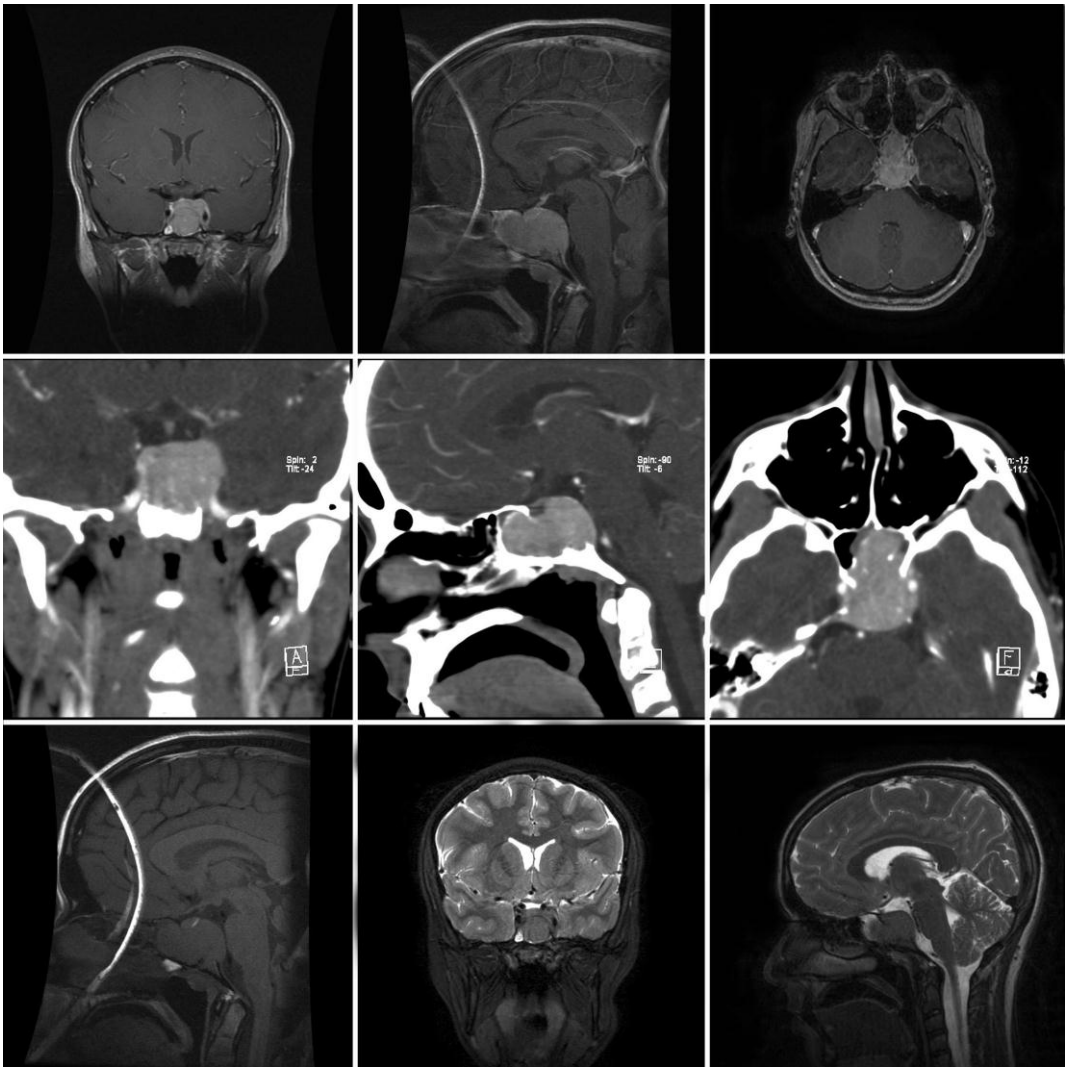

Figure 1: *pre-OP imaging.*

First row:	*MRI T1 with contrast*
Second row:	*cCT with contrast*
Third row:	*native T1- and T2-weighted images*

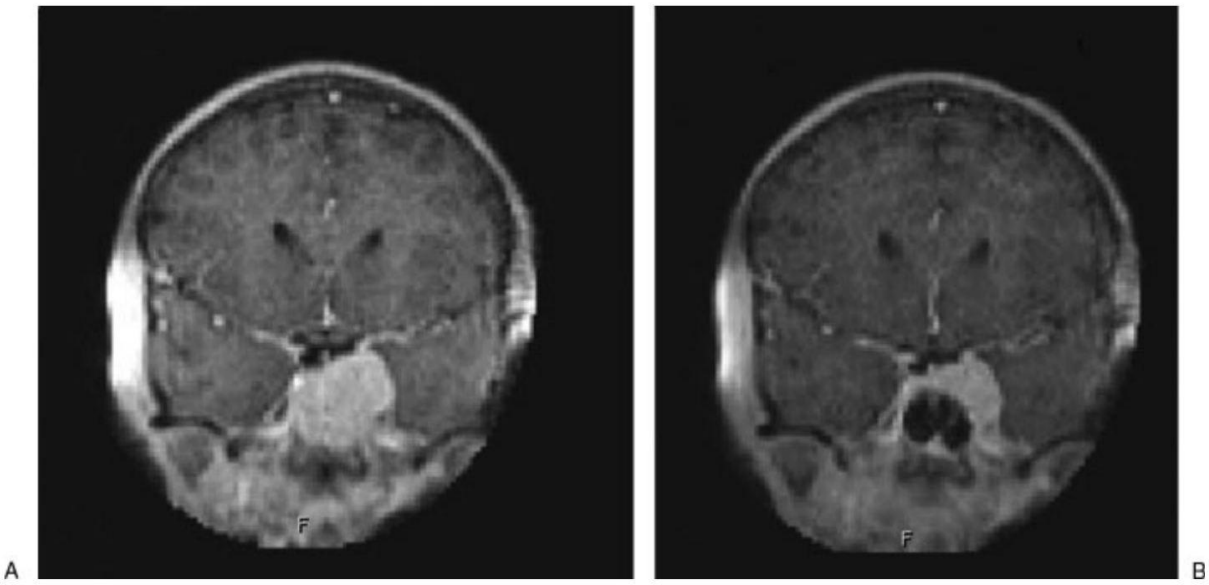

Figure 2: *intraoperative MRI scans (PoleStar N20, 0.15 T, Medtronic Navigation) of the combined procedure.*

a *preoperative T1-weighted scan with contrast* ***b*** *T1-weighted scan with contrast after the transsphenoidal resection*

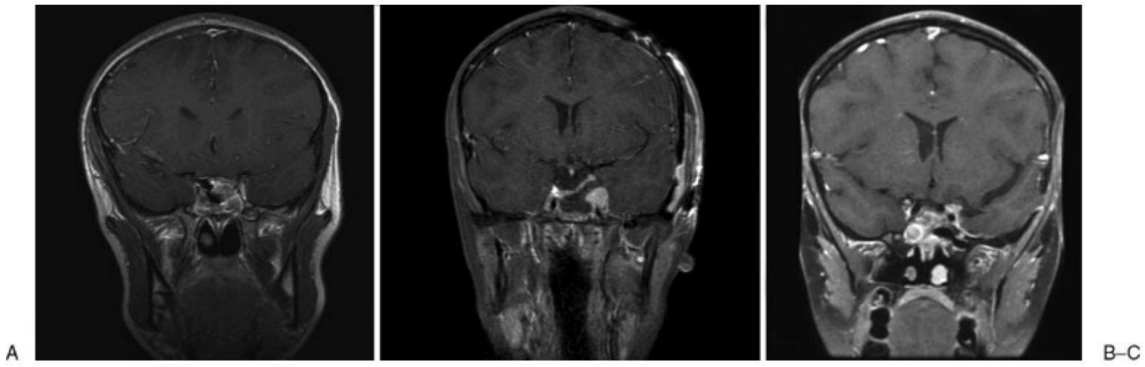

Figure 3: *T1-weighted contrast enhanced follow-up imaging.*

a *4 weeks after the initial transsphenoidal procedure* ***b*** *after the combined approach* ***c*** *after proton therapy and 12 months after the combined approach*

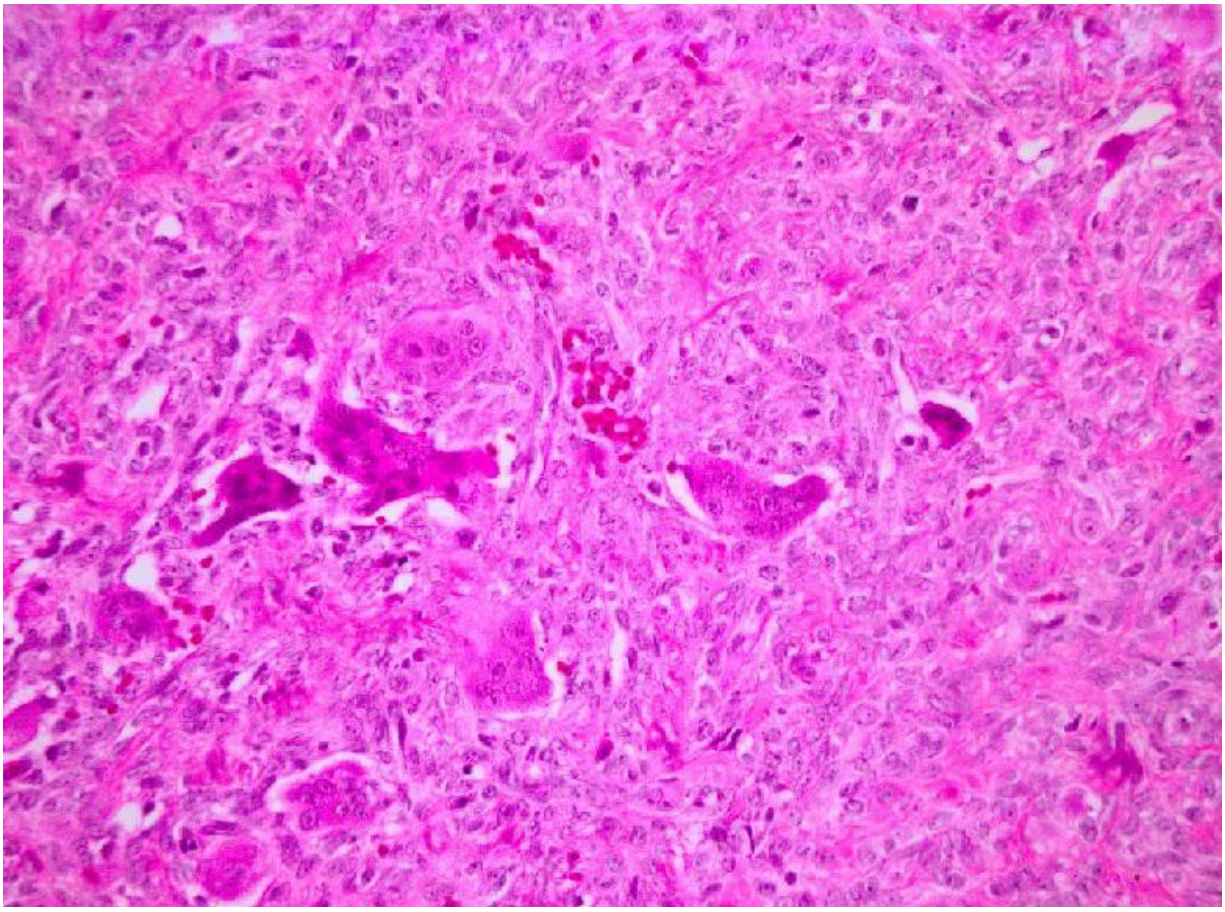

Figure 4: *Microscopic image HE, x20. Cellular lesion containing giant cells and fibroblastic cells in a fibrous, eosinophilic matrix.*

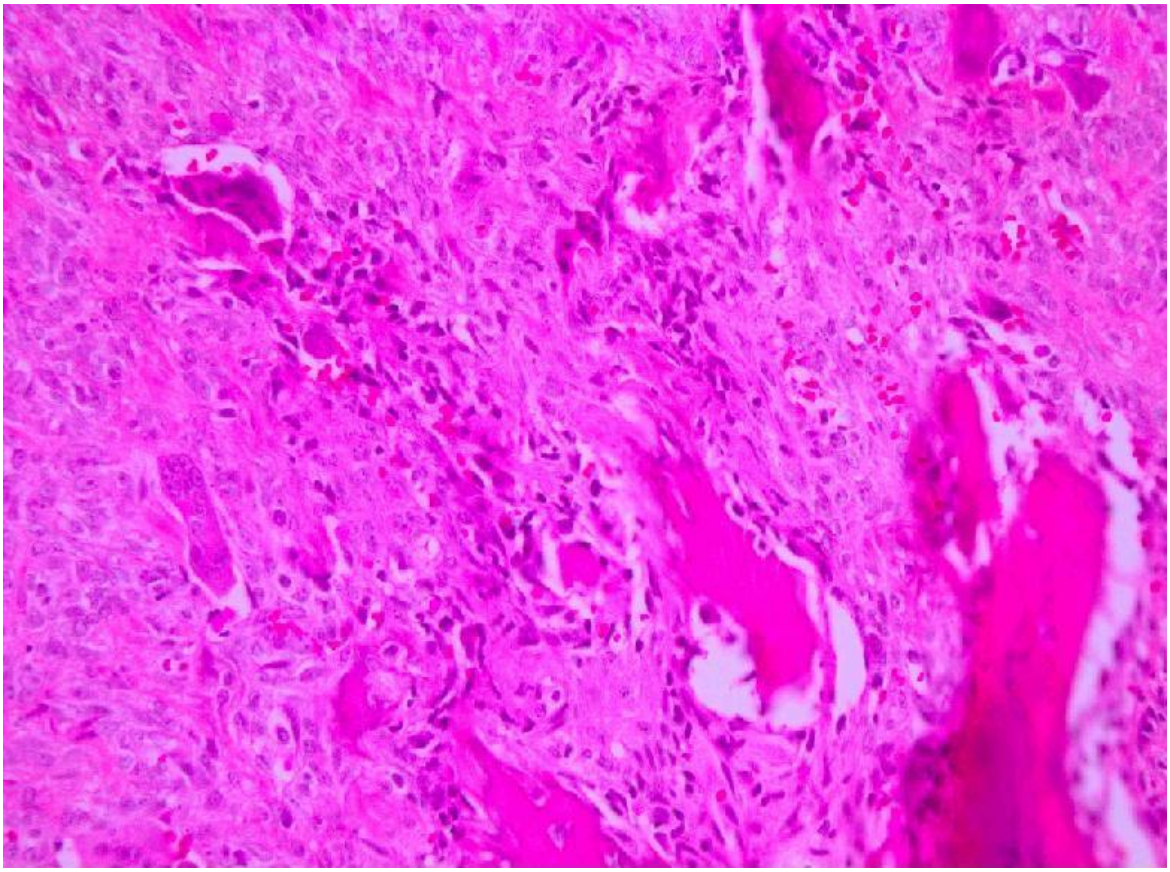

Figure 5: *Microscopic Image HE, x20. The matrix is focally compacted to osteoid, resembling immature bone. At the lower right border, residual non-neoplastic bone can be seen.*

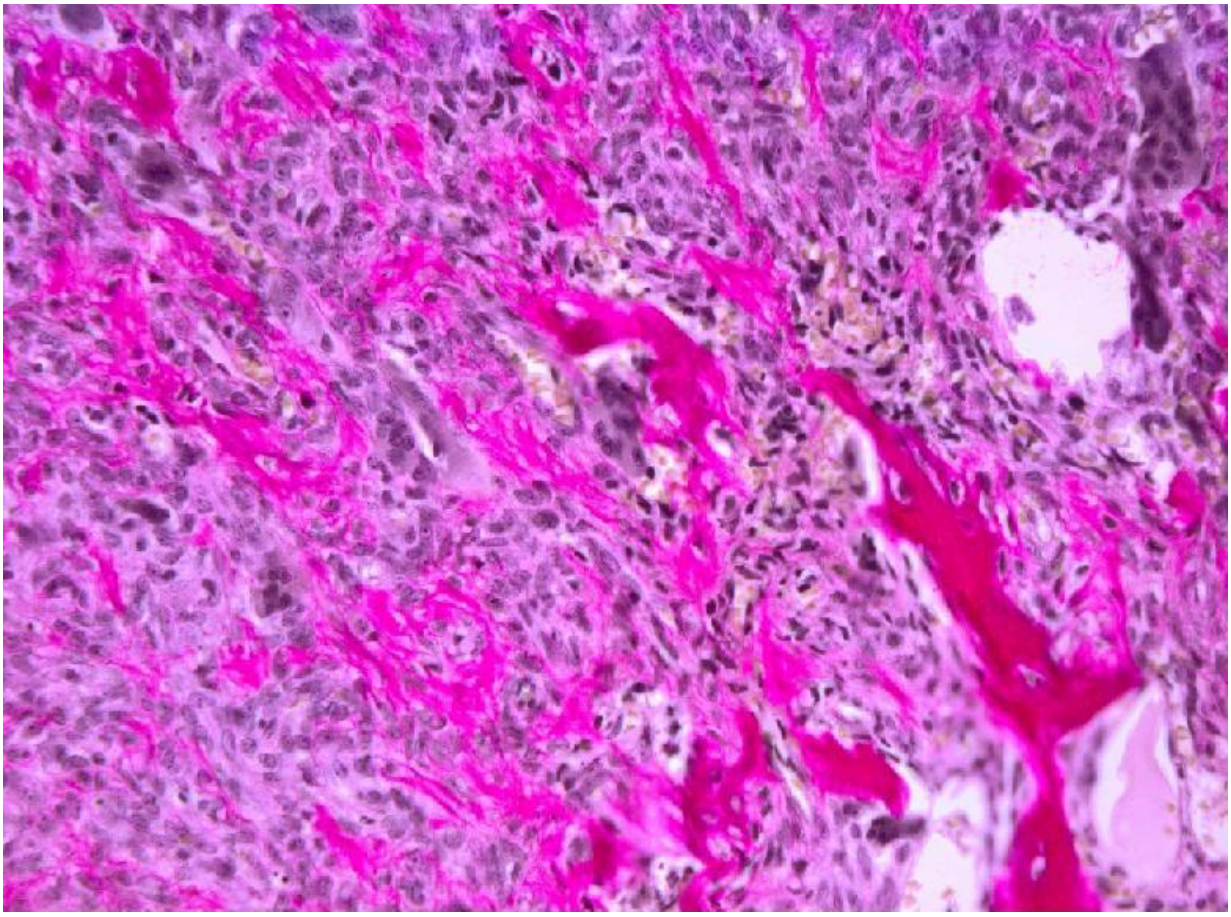

Figure 6: *Microscopic Image EVG, x20. Elastica van Gieson staining reveals a net-like structure of collagen fibers which is compacted to form immature osteoid in some areas.*

Table 1: Endocrinologic data. *the patient received steroid therapy at that time

hormone	pre-OP	post 2nd OP	6 months	12 months	normal range
TSH	1.62 mU/l	1.89 mU/l	2.12 mU/l	3.05 mU/l	0.53 - 3.59 mU/l
fT4	17.6 pmol/l	13.1 pmol/l	14.6 pmol/l	13.4 pmol/l	12.0 - 20.6 pmol/l
LH	10.7 IE/l	7.9 IE/l	11.9 IE/l	7.6 IE/l	depending on age and menstrual cycle
FSH	5.9 IE/l	3.2 IE/l	5.1 IE/l	7.7 IE/l	depending on age and menstrual cycle
prolactin	7.9 µg/l	5.8 µg/l	17.1 µg/l	23.4 µg/l	4.8 – 23.3 µg/l
estradiol	188 pmol/l	85 pmol/l	197 pmol/l	93 pmol/l	depending on age and menstrual cycle
IGF1	293 µg/l	312 µg/l	289 µg/l	177 µg/l	226 – 903 µg/l
cortisol	16 nmol/l *	424 nmol/l	321 nmol/l	608 nmol/l	171 – 536 nmol/l